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Case Report

Multiple papillary fibroelastomas of the aortic valve detected by real time three-dimensional transesophageal echocardiographic images

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ABSTRACT

Papillary fibroelastoma (PFE) is a well-known primary cardiac tumor, but multiple PFEs are rare. We report an interesting case with multiple PFEs that were clearly demonstrated and evaluated with real time three-dimensional (RT3D) transesophageal echocardiography (TEE). A 77-year-old woman was referred to our institution with a diagnosis of osteoarthritis of the hip. Transthoracic echocardiography showed an abnormal structure on the aortic valve. Although two-dimensional TEE revealed typical characteristics of multiple PFE, RT3D TEE clearly demonstrated their number and location on the right and non-coronary cusp of the aortic valve. These results were subsequently confirmed by surgery and pathological findings. RT3D TEE is an exceptionally useful tool for pre-surgical evaluation of PFE.

<**Learning objective:** PFE is a well-known primary cardiac tumor, but multiple PFEs are rare. We report an interesting case with multiple PFEs that was clearly demonstrated and evaluated with RT3D TEE. RT3D TEE is an exceptionally useful tool for pre-surgical evaluation of PFE.>

Introduction

Papillary fibroelastoma (PFE) is the second or third most common primary cardiac tumor and involves both the cardiac valves and chambers [1–5]. The majority of PFEs present as solitary masses, but 5.2% of PFEs present as multiple tumors [2]. Recently, real time three-dimensional (RT3D) echocardiography became available to obtain three-dimensional information on cardiac structure. We report a case with multiple PFEs which was clearly detected by RT3D transesophageal echocardiography (TEE) before surgery.

Case report

A 77-year-old woman was referred to our hospital with a diagnosis of osteoarthritis of the hip. Although she had diabetes mellitus and systemic arterial hypertension, her physical examination was unremarkable, without cardiac murmurs, abnormal heart sounds, or any signs of peripheral embolization. However, electrocardiogram was remarkable for a first degree atrioventricular block with complete left bundle branch block.

Transthoracic echocardiography (TTE) was normal except for mild concentric left ventricular hypertrophy with preserved ejection fraction. TTE also showed an abnormal mobile mass attached to the non-coronary cusp of the aortic valve (Fig. 1). The mass was 10 mm in length and very mobile. Two-dimensional TEE revealed that there were three masses on the cusps of the aortic valve (Fig. 2 and Movie 1).

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Subsequent RT3D TEE with 3D zoom mode (iE33 with X7-2t probe, Philips Medical System, Andover, MA, USA) clearly provided more detailed information on the PFEs – one mobile mass on the right coronary cusp, two on the non-coronary cusp, and none in the left (Fig. 3A and Movie 2). At cardiac surgery, there were two mobile masses on the right coronary cusp, three masses on the non-coronary cusp, and none in the left coronary cusp. The largest at 14 mm was on the non-coronary cusp, and the second largest at

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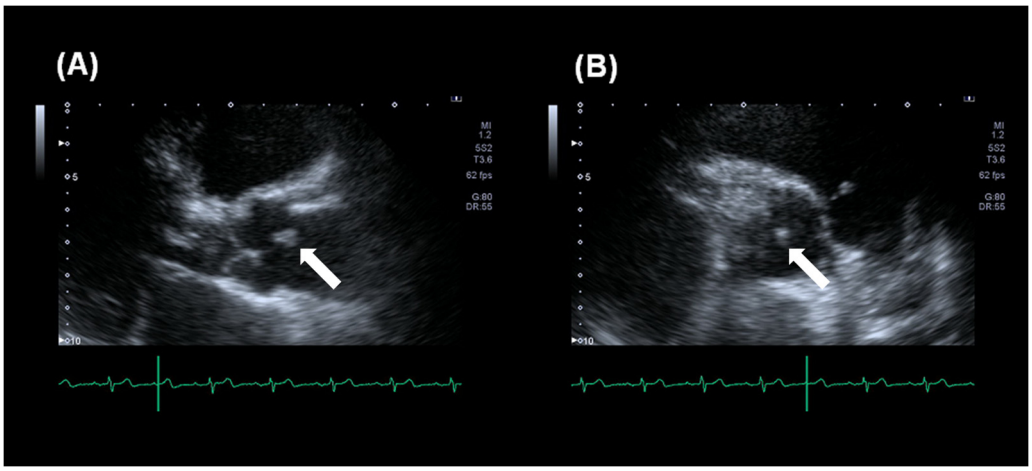

Fig. 1. (A) Parasternal long-axis view in two-dimensional transthoracic echocardiography (magnified at the aortic valve) showing a hyperechogenic mass attached to the aortic valve (arrow). (B) Parasternal short-axis view in transthoracic echocardiography (magnified at the aortic valve) showing a hyperechogenic mass attached to the aortic valve (arrow).

6 mm was also on the non-coronary cusp (Fig. 4A). The other two masses that RT3D could not detect were less than 2 mm in length. The surgery was successful for shave excision of tumor and preserved native aortic valve without any residual aortic valve function abnormality. Pathological findings were consistent with those of PFE (Fig. 4B).

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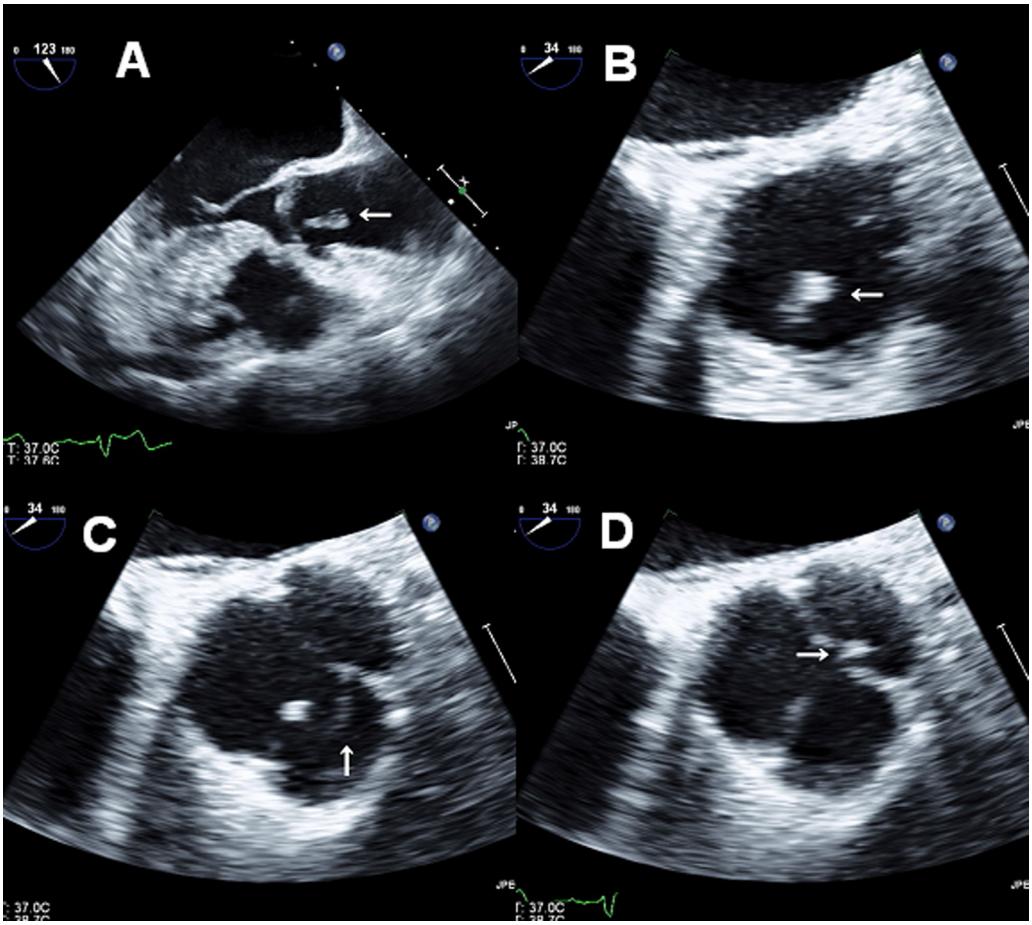

Fig. 2. (A) Two-dimensional transesophageal long-axis view showing a hyperechoic mass attached to the non-coronary cusp of the aortic valve (arrow). (B) Transesophageal short-axis view (magnified at the aortic valve) showing a hyperechoic mass attached to the non-coronary cusp of the aortic valve (arrow). (C) Transesophageal short-axis view (magnified at the aortic valve) showing a hypoechoic mass attached to the right coronary cusp of the aortic valve (arrow). (D) Transesophageal short-axis view (magnified at aortic valve) showing a hyperechoic mass attached to the left coronary cusp of the aortic valve (arrow).

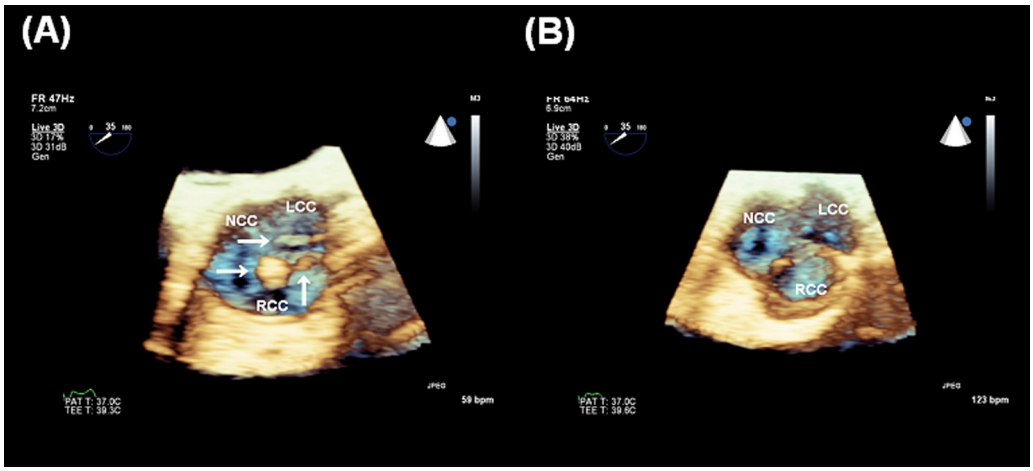

Fig. 3. (A) Aortic valve image by three dimensional zoom transesophageal echocardiography (TEE) showing three masses attached to the non- and right coronary cusp and of the aortic valve (arrow). (B) Aortic valve image by three dimensional zoom TEE after removal of papillary fibroelastoma. RCC, right coronary cusp; LCC, left coronary cusp; NCC, non-coronary cusp.

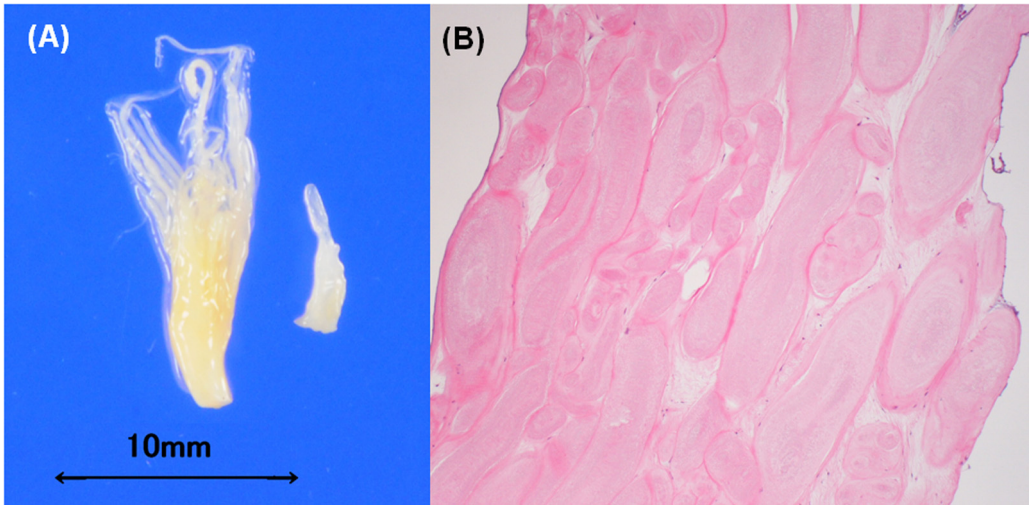

Fig. 4. (A) Gross picture of the resected papillary fibroelastoma attached to the non-coronary cusp. (B) Hematoxylin-and-eosin staining shows that revealing typical papillary architecture of cardiac papillary fibroelastoma (papillary fronds lined by a single layer of endothelium overlaying hyalinized stromal cores). Original magnification 100×.

Discussion

The term "papillary fibroelastoma" was introduced in 1975 by Cheitlin et al. [6]. PFE is the second or third most common primary cardiac tumor and involves both the cardiac valves and chambers [1–5]. The majority of PFEs present as solitary masses, but 5.2% of PFEs present as multiple tumors [2].

Although PFE was considered until recently to be benign, there are several reports that it is associated with critical complications, such as coronary ostial embolism [7] and acts as a substrate for fibrin and platelet aggregation which leads to peripheral and central embolization [3]. Therefore, a clinical decision was made to remove these tumors. There is some controversy about PFE management. However, mobile PFEs and/or PFEs larger than 10 mm were recommended for surgical removal even when a patient was asymptomatic [8] because of PFEs' potential complication risk and low surgical risk. More than 80% were successfully removed by a simple excision [2]. RT3D TEE before discharge confirmed normal native aortic valve structure and function (Fig. 3B).

There are several reports of multiple PFEs on the aortic valve that were diagnosed by two-dimensional TEE [9–11] and 3D TTE [12]. However, 3D TEE can provide better quality images than two-dimensional TEE or 3D TTE. This is the first report that multiple PFEs were examined, diagnosed, and precisely described by RT3D TEE. Because RT3D TEE enables us to assess the 3D structure of the heart, it is useful for finding and evaluating the exact location and mobility status of PFEs, especially when multiple. It is not an easy task for clinicians to recreate precise 3D structures from two-dimensional images in their minds. Surgery revealed five masses on the aortic valve, but 3D TEE detected only three. The other two masses were <2 mm in length and undetected by RT3D because extremely small PFEs are difficult to identify by echocardiography [4].

Conclusion

This is the first report of multiple PFEs on the aortic valve documented using RT3D TEE which demonstrates that RT3D TEE is an exceptionally useful tool for pre-surgical evaluation of PFE.

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